SKIN MANIFESTATIONS OF INTERNAL DISEASE

A TALE OF TWO SYNDROMES

Gopikrishnan Anjaneyan [1] - Mithun Cb [2]

Amrita Institute Of Medical Sciences, Dermatology, Kochi, India [1] - Amrita Institute Of Medical Sciences, Rheumatology, Kochi, India [2]

Background: Pancreatic panniculitis is an uncommon condition affecting 2% of patients with pancreatic disease. When polyarthritis is also present it's an even rarer entity called pancreatitis-panniculitis-polyarthritis(PPP) syndrome. Pancreatic enzymes amylase and lipase are released in to systemic circulation leading to lipolysis, secondary inflammation of subcutaneous tissue and joints.

Observation: A 29 year old male presented with multiple painful red nodules over both legs for 4months. He had a past history of vomiting and abdominal pain 6 months back and was being evaluated for non-alcoholic pancreatitis.

Differentials included erythema nodosum, erythema induratum, pancreatic panniculitis and polyarteritis nodosa.
5 days later,he developed painful swelling of bilateral knee, ankle and wrists, along with fever and multiple fresh large tender subcutaneous nodules over trunk and extremities.

Investigations showed raised ESR(30),CRP(188),Serum lipase(11960U/L),amylase(4754U/L). Mantoux test, blood and pus cultures and were negative. RA factor, ANA,ANCAs, complements C3,C4, HepatitisB&C were negative. X-ray showed soft tissue swelling of involved joints. Arthrocentesis demonstrated yellowish fluid, which was culture negative(excluding septic arthritis). MRI Abdomen showed well defined T2 hyperintense foci measuring 1.4x1.5cm over head of pancreas. Skin nodule histopathology demonstrated subcutaneous fat necrosis & foamy cell change and "ghost like fat cells".

Panniculitis on histopathology, investigations confirming pancreatic disease and reactive arthritis confirmed Pancreatitis-Panniculitis-Polyarthritis(PPP) syndrome.
Within 4 days his clinical & laboratory indicators worsened -High ferritin(2948ng/ml), CRP(248mg/ml) ,SGOT (51u/L), Amylase (11450u/L) and lipase (5640u/L) low platelet count(58,000)and this patient developed Macrophage Activation syndrome(MAS). He was managed with appropriate medications under rheumatology and dermatology departments but despite all efforts he succumbed.

Key message: Pancreatitis-Panniculitis-Polyarthritis(PPP) Syndrome is extremely rare and

only around 30 cases are reported in literature. There is no current reference in literature of MAS coexisting with PPP syndrome. Management should be mainly directed towards treating underlying pancreatitis and early recognition is indispensable owing to high mortality associated with it.